

NEUROFIBROMA OF THE LIP- REPORT OF A RARE CASE.

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ABSTRACT

Neurofibroma is a rare tumor of the oral cavity. It is generally associated with a generalized syndrome of neurofibromatosis but a few cases of the solitary intraoral lesions have also been reported. A one such rare case of neurofibroma of the lip is being presented with a note on the importance of early diagnosis and adequate treatment of this lesion.

KEY WORDS : solitary, neurofibroma, neurofibromatosis

INTRODUCTION

Neurofibromata are uncommon tumors of the oral cavity. Although they represent one of the common entities among neurogenic tumors, its occurrence as an intraoral tumor is uncommon.¹ They are predominantly seen as a feature associated with the generalized syndrome neurofibromatosis (also called von recklinghausen's disease, named after the German pathologist who first documented its features), but a few cases of solitary neurofibromas of the jaws have also been reported.²⁻⁴ Solitary neurofibroma of the jaws was first reported by Bruce in 1954⁵ after which not many cases have been reported in literature.

Case report:

A 54 year old male patient reported to our unit with a complaint of a painless swelling on the left side of his lower lip. The swelling had started about 40 years back and grew slowly to attain its present size. On examination, there was an obvious enlargement of the left side of his lower lip extending from the left commissure and crossing the midline. A well circumscribed 2x3cm lesion was seen on the left labial mucosa not extending into the vestibule. The mucosa over the lesion was normal with no redness or draining sinuses seen over it. On palpation, the swelling was non-tender, firm in consistency, nodular and fixed to the underlying tissues. The lesion was non- pulsatile and non

reducible. There was no altered sensation on the lower lip.

The patient was moderately built and nourished and did not have any other swellings palpable elsewhere in the body. Careful examination was done to rule out Lisch's nodules and Crowe's sign.

A provisional diagnosis of a benign tumor was made and an excisional biopsy was planned under local anesthesia.

A bilateral mental nerve block was given and a superficial mucosal incision was made over the lesion. A large mass was seen to emerge from underlying lesion. The growth was carefully dissected and separated from the underlying tissues to reveal a firm, whitish coiled "worm-like" structure. The excised specimen was almost 7cm in length. It was sent for histopathological examination. The bed of the lesion was examined for remnants, hemostasis achieved and sutured. The post operative course was uneventful. The patient was kept on a constant follow up and was seen to be disease free 3 years after the surgical excision with no signs of recurrence.

The histopathology report showed the tumor to be composed of proliferative, elongated spindle shaped cells arranged in fascicles with dark stained



Fig.1. Lower lip swelling extending from the commissure and crossing the midline



Fig 2. Intraoral enlargement seen on the labial mucosa



Fig.3. coiled tumor emerging from the bed of the lesion



Fig.4. Excision of the tumor

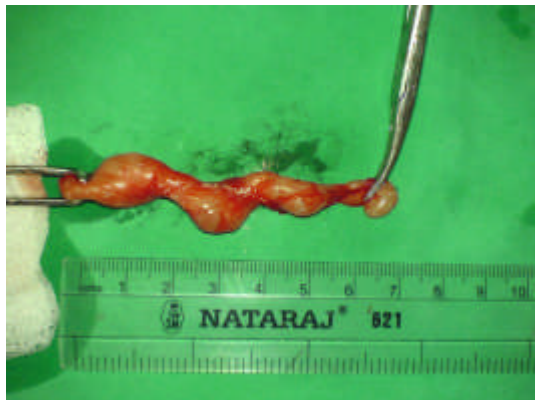


Fig.5: excised specimen

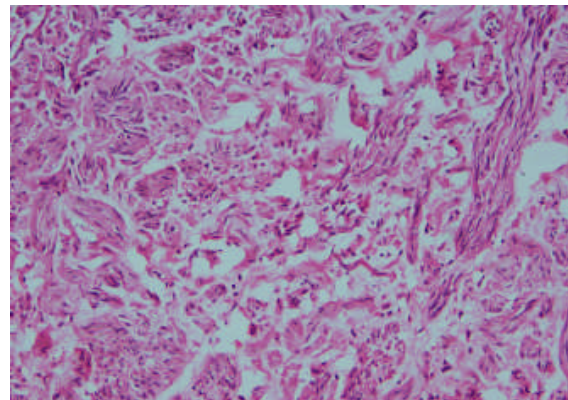


Fig 6: photomicrograph showing elongated spindle shaped cells with dark stained nuclei suggestive of cellular type of neurofibroma

nuclei. The stroma was composed of collagen fibres and some mucoid material. Silver stain was used to demonstrate the presence of axons in the tissue. This confirmed the diagnosis of a solitary neurofibroma (cellular variety) of the lip.

Discussion

Neurofibroma of the oral cavity is a rare, benign, non-odontogenic tumor. Many different forms of neurofibroma have been described in literature ⁽⁶⁾. These include:

- Cutaneous neurofibroma (localized and diffuse)
- Intraneural neurofibroma (localized and plexiform)
- Massive soft tissue neurofibroma (diffuse and plexiform)
- Visceral neurofibroma(solitary or multiple)
- Sporadic or associated with neurofibromatosis type I (von Recklinghausen's disease)

Although all these entities have common histopathological features they differ mainly on their clinical presentation and the gross appearance.

Neurofibromatosis is an autosomal dominant disease which affects the neural crest cells that give rise to ectodermal and mesodermal derivatives. This genetic disorder affects 1 in every 3000 of the population and has the highest mutation rate among genetic disorders.⁷ Only few cases of solitary neurofibroma have been reported in the literature^{8,9}

Von Recklinghausen's disease which occurs as a result of an abnormality of chromosome 17 is described to have the characteristic features which include Cafeaulait spots, multiple neurofibromas, Lisch nodules (hamartomas of the iris) and Crowe's sign (axillary and inguinal freckling). Only few cases of solitary neurofibromas have been reported in literature. Solitary neurofibroma, by definition is seen in those patients who do not have neurofibromatosis.^{10,11}

The common sites of occurrence intraorally include the tongue. Other sites reported are the floor of the mouth, buccal mucosa, edentulous alveolar ridge and the pharyngomaxillary spaces.⁷ Around 40 intraosseous solitary neurofibromas have been reported in literature. Most of the intraosseous varieties are present in the posterior mandible with only few of the reported cases being in the maxilla.

There is a female predilection in the ratio of 2:1 ^(12,13). These tumors generally occur in the 3rd decade but have been reported to occur between the ages of 10 months to 70 years.

These lesions are described as pedunculated or sessile nodular growths. They are always slow growing and mostly painless growths. Pain and paresthesia may be due to nerve compression.^{14,15} Some cases where the lesion occurs in the tonsillar region, the patient may complain of dysphagia.

The excised specimen appears as a firm whitish mass which has a shiny surface. Microscopically, the tumor is composed of interlacing proliferating bundles of elongated spindle shaped cells with dark stained nuclei. The stroma is composed of thin collagen fibres and mucoid material. Silver staining may be useful in demonstrating small axons in the tumor tissue.¹⁶ In almost 85-100% of the cases immunohistochemistry stains positive for S-100 which confirms its neural tissue origin.^{17,18}

A solitary neurofibroma must be differentiated from a schwannoma. A schwannoma is encapsulated, eccentric to the nerve and composed of Schwann cells. A neurofibroma on the other hand, incorporates the nerve (which may or may not be identifiable) and it is composed of Schwann cells, perineural-like cells, fibroblasts and transitional cells.⁶

Over time, neurofibromas have the propensity for progression into neurofibromatosis. Although these are originally benign lesions they also have a likelihood for malignant transformation. Almost 6-29% of malignant transformation has been reported.^{19,20}

Surgical excision by conserving the nerve of origin is the treatment of choice.³ In many cases it may be impossible to separate the tumor completely from the nerve in which case it may be advisable to resect the nerve also. Some cases of spontaneous remission of the tumor after puberty have been reported and some authors have suggested surgical excision after puberty.²¹ However, to prevent excessive disfigurement and neural deficit it may be advisable to excise the tumor on detection. There have been variable reports regarding the recurrence of this lesion. Although some studies report recurrences as rare some report a significantly high rate of recurrence.^{22,23} Our patient was seen to be disease free three years after surgical excision.

Although these are benign tumors and have a fairly good prognosis, since they have a propensity for malignant transformation and also chances of progressing into neurofibromatosis, these lesions must be monitored carefully and treated meticulously. Even though they are a rare lesion in the oral cavity, solitary neurofibromas must be considered in the list for differential diagnoses in cases of intraoral swellings and intraosseous lesions of the jaws.

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